Granulosis rubra nasi

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ABSTRACT

Granulosis rubra nasi is a rare disorder of the eccrine glands, inherited as an autosomal dominant trait. It is clinically characterized by hyperhidrosis of the central part of the face, most conspicuous on the tip of the nose, followed by appearance of diffuse erythema over the nose, cheeks, chin, and upper lip. It is commonly seen in childhood, but can also occur in adults. This is a case report of a 27-year-old male patient who presented with excessive sweating over the nose. Physical examination of the nose revealed erythema and multiple telangiectatic vesicles. Biopsy findings supported the diagnosis of granulosis rubra nasi. This case is being reported for its rarity since to the best of our knowledge, it has not been reported in Indian subjects so far.

Key words: Disorder of eccrine glands, granulosis rubra nasi, telengiectasia

INTRODUCTION

Granulosis rubra nasi is a rare disorder of the eccrine glands first described by Jadassohn in 1901. This disease is also known as "Acne papulo-rosacea of the nose."[1] It is thought to be common in European countries. In the English literature Maschkillesson and Neradow (1935) have recorded 130 cases and the disease was not thought to be rare.[2] Although it is inherited as an autosomal dominant trait, it is rare to find the disease in two generations, although less commonly it is found in two children of same parents.[2,3] The disease generally appears in the childhood but adolescent or adult onset is possible.[3] Granulosis rubra nasi is a focal form of hyperhidrosis that differs from the other forms in that it does not depend on the hypothalamic or emotional stimuli to develop.[1]

CASE REPORT

A 27-year-old man presented with erythema over the nose since childhood without sensitivity to sunlight. He was otherwise well and there was no relevant family history. Physical examination showed excessive sweating of the nose, erythema of the nose covered by beads of sweat, and multiple telangiectatic vesicles [Figures 1 and 2]. The rest of the physical examination was normal. The skin biopsy showed dilatation of blood vessels, dilated sweat ducts with a discrete mononuclear cell infiltrate surrounding them [Figure 3]. The patient was reassured taking into consideration the benign nature of the disease.

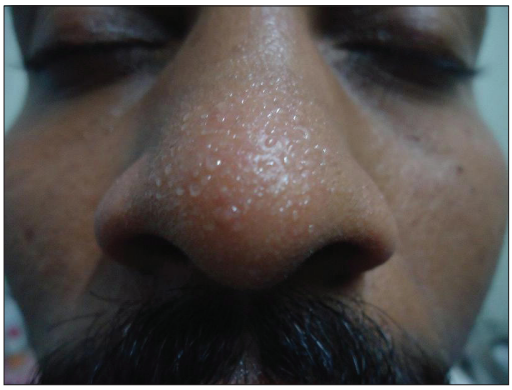

Figure 1: Beads of sweat seen on an erythematous base over the nose

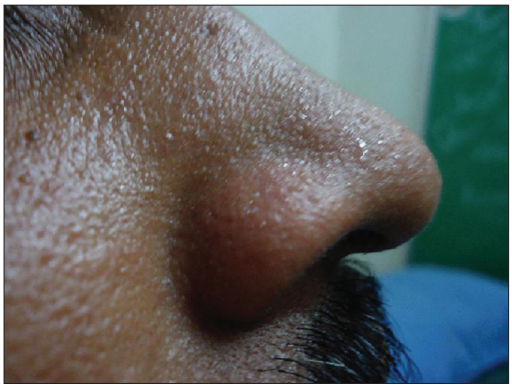

Figure 2: Shows erythema more prominently. Telangiectasias are appreciable

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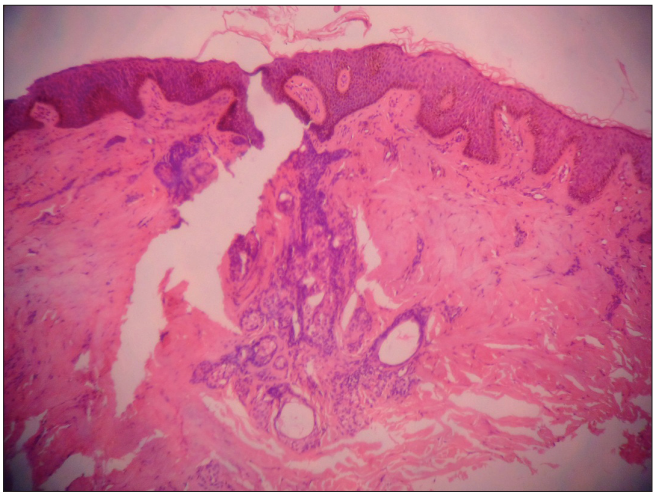

Figure 3: Histopathology section showing dilated blood vessels and eccrine ducts with surrounding lymphocytic infiltrate [H & E, 10X]

DISCUSSION

Our patient presented with granulosis rubra nasi, a rare disease. It usually starts in childhood in patients aged between 6 months and 15 years. The clinical picture is characterized by hyperhidrosis of the central part of the face, most conspicuous on the tip of the nose. This is followed by appearance of diffuse erythema over the nose, cheeks, chin, and upper lip. Erythematous macules, papules, vesicles or even pustules may be seen over the sweat duct orifices.[4] Later numerous small telangiectasias may occur over the nose, cheeks, and chin.[1,5] Comedo-like lesions may also be present.[1] The nose may be palpably cold and cystic lesions can develop.[3] It may sometimes be associated with acrocyanosis and hyperhidrosis of the palms and soles. The diagnosis is largely clinical. Histopathology shows a mild mononuclear cell infiltrate around the sweat ducts, blood vessels, and lymphatics. There is dilation of blood vessels and the lymphatic vessels. Pilosebaceous units are normal and no heterotopic apocrine glands are found[3] This disease usually remits after puberty, unlike the primary forms of localized hyperhidrosis but sometimes may continue into adulthood.[3,4] There is neither systemic associations nor abnormal laboratory findings.[3]

The pathogenesis of the disease is still unknown. Some authors have suggested that it is a disorder of vasomotor and secretory function of the nose. Goldsmith had described rhino rrhea as an associated feature.[6] Eddowes had suggested that adenoids could be involved which can provide a source of irritation to the tip of the nose.[7] Heid *et al*. reported a case of granulosis rubra nasi associated with phaeochromocytoma, surgical removal of which was followed by regression of the condition.[8]

The clinical picture is very distinctive and rarely there is a problem with the diagnosis. The differential diagnosis includes rosacea and perioral dermatitis. In rosacea there is erythema of the cheeks and nose along with telangiectasias but no hyperhidrosis of the central part of the face. In the case of perioral dermatitis there are small monomorphic papules, pustules, erythema, and scaling involving the perioral area and no hyperhidrosis. Other differential diagnosis include acne vulgaris, lupus pernio, and lupus erythematosus.[3] Treatment of granulosis rubra nasi has been described in the literature with topical indomethacin, oral corticosteroids, tetracycline, cryotherapy, and even X-rays.[2] However, reassurance is more important. Drying lotions like calamine can be tried.[3] Grazziotin *et al*. have recently described a treatment using botulinum toxin A that induced long-term remission in a patient with granulosis rubra nasi.[9]

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